

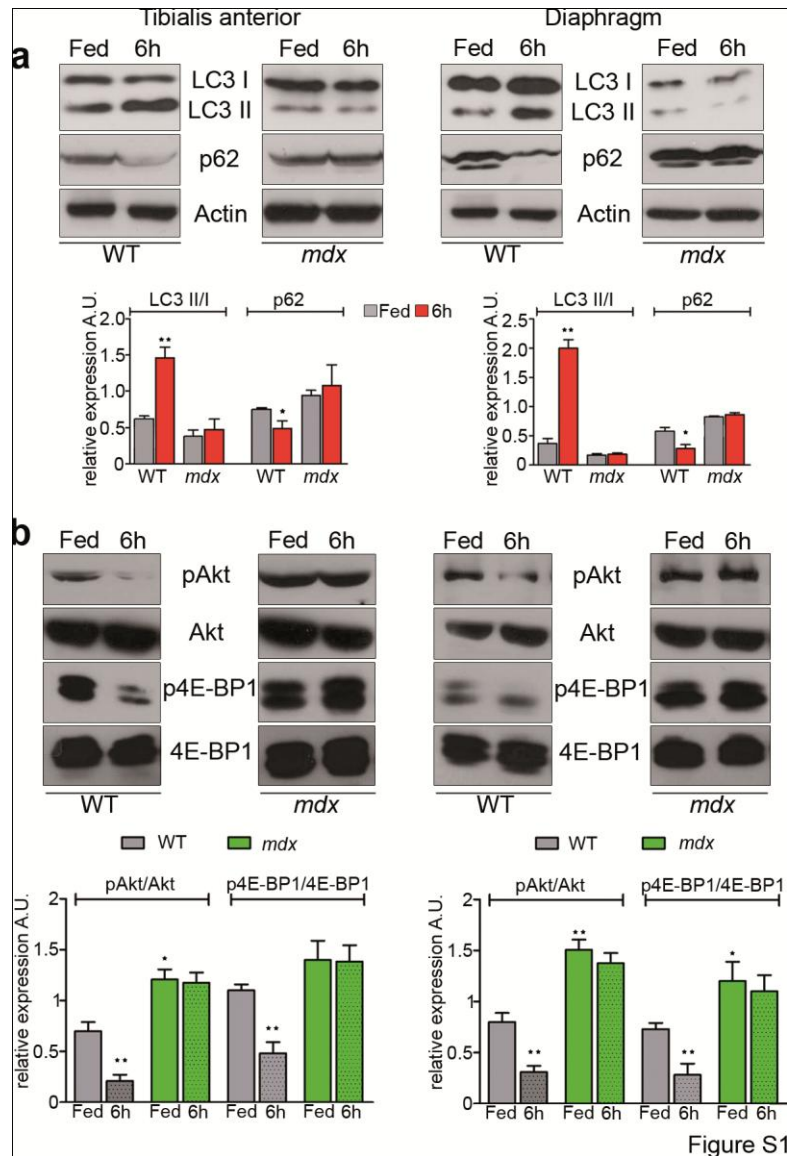
SUPPLEMENTAL MATERIAL

Autophagy as a new therapeutic target in Duchenne muscular dystrophy

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SUPPLEMENTAL FIGURES

Supplemental Figure 1

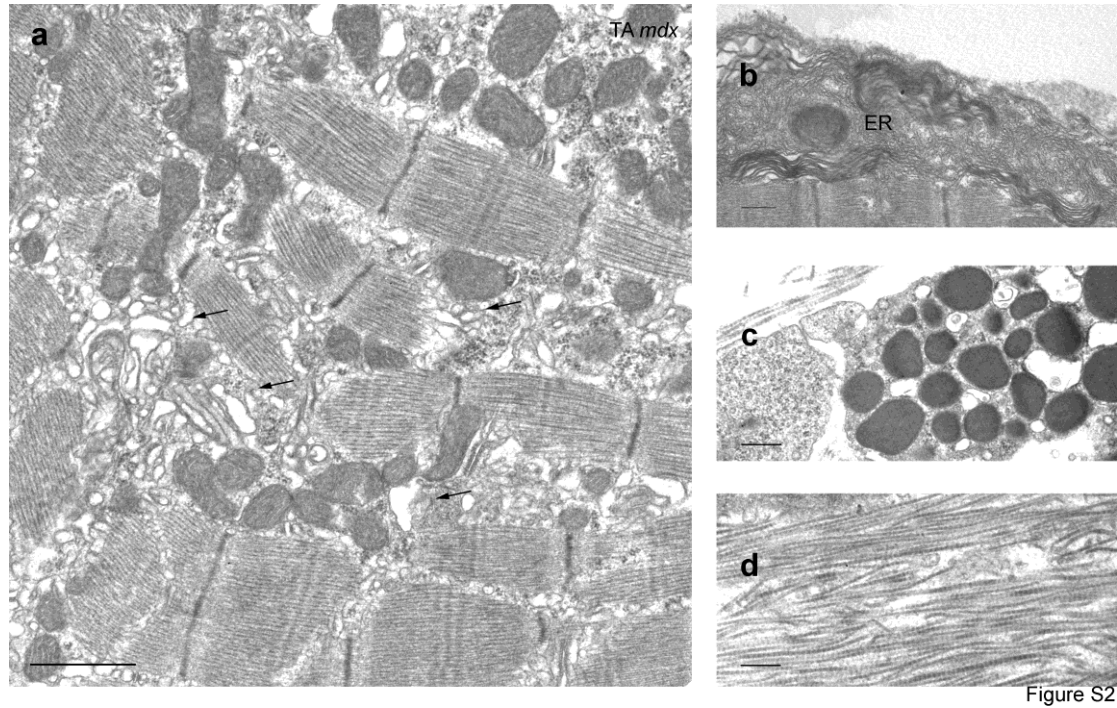


Supplemental Figure 1 Autophagy induction is impaired in *mdx* mice. **(a)** Western blots showing LC3 lipidation (LC3 II) and p62 levels in *Tibialis anterior* and diaphragm muscles (60 μ g/lane) of fed and 6 h-starved (6 h) WT and *mdx* mice. Densitometric quantification of LC3 ratio and p62 levels, normalised against actin, shows induction of autophagy only in WT mice after 6 h of starvation. The values are

average of two experiments with five animals per group and error band indicate SEM.

(b) Western blots revealing reduction of Akt and 4E-BP1 phosphorylation in *Tibialis anterior* and diaphragm muscles (60 µg/lane) of fed and 6 h-starved (6 h) WT and *mdx* mice. *mdx* muscles display no changes in the phosphorylation levels of both proteins. Western blots in a and b are representative and quantifications correspond to 10 animals per group. Asterisks indicate statistical significance *vs.* WT mice (*P < 0.05, and **P<0.01). Error bars represent SEM.

Supplemental Figure 2



Supplemental Figure 2 *mdx* mice display accumulation of aberrant organelles. **(a)** electron micrographs of *Tibialis anterior* muscles from *mdx* mice. Swollen stacks of ER *cisternae* are present in the intermyofibrillar space. **(b)** ER stacks are packaged in complex structures in the subsarcolemmal region. **(c, d)** infiltrating cells and collagen are abundant in the intermyofibrillar space (scale bars: 500 nm). Images are representative of reproducible results in 10 animals per group.